Aus dem Universitäts-Institut für Röntgendiagnostik und Strahlentherapie
Bürgerspital Basel
Vorsteher: Prof. Dr. E. ZDANSKY
Arbeit unter der Leitung von Prof. ZDANSKY

Constrictive Pericarditis; a Summary of the Disease, and the Role of Roentgenology in its Detection

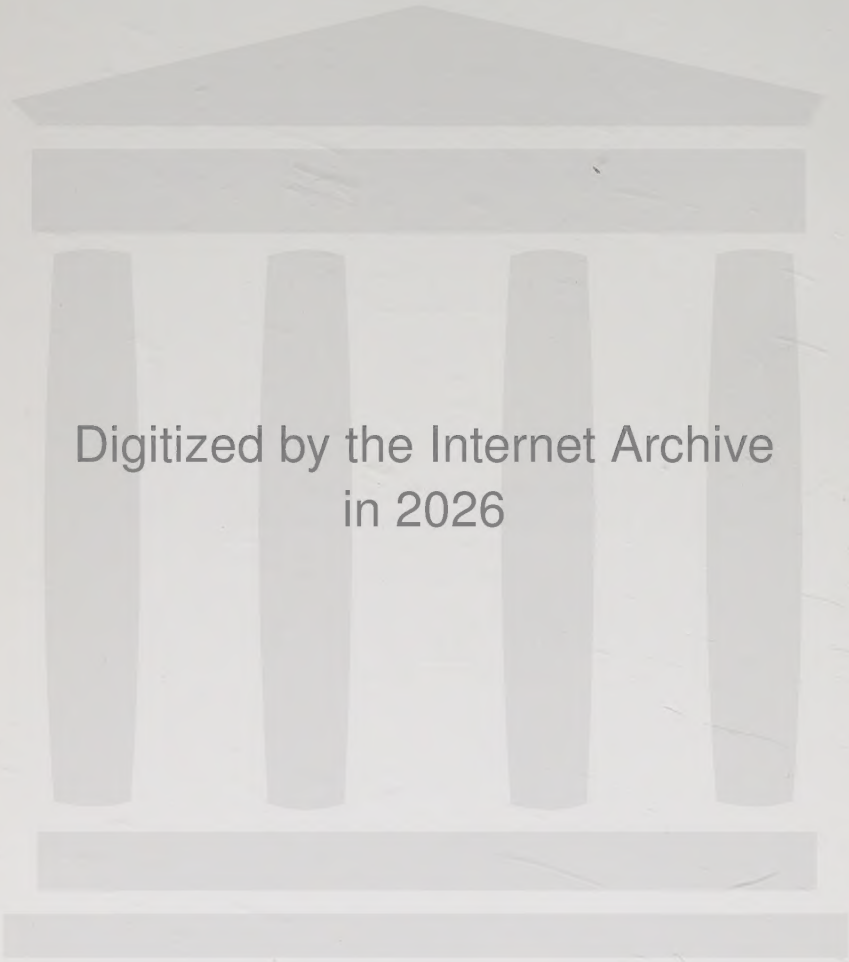
INAUGURALDISSERTATION

zur Erlangung der Doktorwürde
der medizinischen Fakultät der Universität Basel

vorgelegt von

HARRY WALDMANN
von New York (USA)

Basel
Buchdruckerei Birkhäuser AG.
1962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As late as 1883 the celebrated surgeon Billroth stated that 'the surgeon who would attempt to suture a wound of the heart would lose the respect of his colleagues'. With the rapid advance in surgical techniques today, many heart ailments that were viewed as nothing more than pathological curiosities less than sixty years ago are being successfully operated upon. Constrictive pericarditis is such a disease and therefore a review of the malady as well as the techniques available for its early recognition is essential. Early recognition is particularly important because of the very nature of the process. Without treatment the pericardium slowly shrinks and stifles the heart action. Furthermore, it must be pointed out that the longer the disease remains untreated, the greater becomes the likelihood that myocardial atrophy and hepatic failure, due to stasis, will reach an irreversible stage. An operation performed at this time will not bestow lasting benefit. MORTENSEN and WARBURG (1948) calculated the life expectancy of an untreated patient to be 6 to 13 years from the onset of symptoms. It was observed that if constrictive pericarditis commenced before puberty bodily development was arrested, but the loss was quickly compensated post-operatively. The earlier the diagnosis is made the smaller is the surgical risk to the patient and the greater his chance for a complication-free post-operative course.

On section the pericardium may appear as a mass of dense, fibrous or fibro-calcific scar tissue firmly connected with the heart muscle. The scar may vary from 0.5 cm to over 1 cm in thickness and varying amounts of calcium or even bone may be found in the inner layer of the scar formation. It frequently invades the myocardium itself and if the entire heart is encapsulated in calcific plates, the term 'armored heart' is employed. The heart itself is small or normal in size, weighing 175 to 300 gms. VOLHARD (1923) differentiates an internal and external type of constrictive pericarditis. The internal type (Concretio cordis) is characterized by a thick adherent pericardium whereas the external (Accretio cordis) shows adhesions extending to the endothoracic fascia and mediastinal pleura thereby cementing the pericardium to the anterior chest wall, mediastinum, spine or diaphragm. Histologically, the scar shows acellular fibrosis with some hyalinization consistent with chronic inflamation. Very often fibroblastic proliferation, vascularity and inflammatory cells can be seen. The heart itself often shows signs of atrophy. ROBERTS and BECK (1941) examined the hearts of three patients who died after being operated on for constrictive pericarditis. The transverse diameter of cut heart muscle was reduced by 30% of the normal measurement. These writers produced cardiac constriction in dogs and also found 30% reduction in muscle fibers. In the human hearts the lack of fibers of varying thickness, typical of atrophy, was also noted.

Chronic passive hyperemia of the lungs or in the systemic circulation is frequently seen at autopsy. Whether the pulmonary or systemic circulation will show signs of congestion is dependent on the location of the constricting process in either the right or left side of the heart.

Although the pathological-anatomic picture presented by pericardium removed at operation or autopsy is very distinctive, it offers practically no help in the determination of the origin of constrictive pericarditis. Usually the fully developed syndrome is a residual state following a healed infection. The scar tissue masks the causative factor so well that in most cases there is no evidence pointing to a specific disease process. Under such circumstances it is only natural that there should be a diversity of opinions concerning etiology. The following examples from the literature should serve to point out the various opinions expressed. Of 28 patients with constrictive pericarditis examined by BLALOCK and BURWELL (1941) 18 were reported to be of tubercular origin, and in 3 cases staphylococcus aureus was held responsible. In many others the impression was gained that tuberculosis was the most probable causative agent. In a very comprehensive study of 61 cases CHAMBLISS *et. al* (1951) classified 28.2 % as tuberculous, 16.4 % as having an acute onset with infection ($^1/_2$ of these being upper respiratory infections), and 4.9 % as acute nonspecific pericarditis. 57.7 % had to be classified as unknown etiology.

HARVEY and WHITEHALL (1937) reviewed 95 cases of tuberculosis of the pericardium and concluded: '. . . we have never seen a patient who recovered from the active process develop chronic constrictive pericarditis'.

In the above mentioned series by CHAMBLISS *et al.*, 10 patients had a past history of rheumatic fever but this could be considered as a possible aetiology in only one case. In following up 1500 cases of rheumatic heart disease HARRISON and WHITE (1942) found no cases of chronic compression.

Individual cases, following trauma, tularemia and metastatic carcinoma have been reported but they are rare.

Most authors believe tuberculosis to be the most common cause since chronic inflammatory changes, areas of caseation and calcification resembling healed tuberculous foci elsewhere in the body can be seen histologically. However, it must be pointed out that all necrotic tissue which cannot be absorbed tends to calcify and therefore it should be possible for pericarditis of any origin to terminate in constrictive pericarditis. Traumatic hemorrhage into the pericardium can organize and thereby cause the same end result. A large number of cases, however, show no history of bacterial or traumatic origin and must be classified as acute, nonspecific or idiopathic pericarditis.

The stifling effect of the pericardium on the underlying heart manifests itself first in a reduction of diastolic filling and, as the pericardium becomes thickened and calcified, in faulty systolic emptying. Which of the two factors is more important in the pathological physiology of the disease is a moot point. Their combined result is a diminished stroke volume evidenced by stasis in the systemic veins and, with a large degree of variability, in the pulmonary circulation. According to Starling's law, cardiac output increases in a direct relation to increased length of cardiac muscle fibers being stretched during diastolic filling. This mechanism is lost in constrictive pericarditis and the regulatory mechanism of circulation tries to compensate this by an increased atrial pressure. Since the cardiac output cannot be raised substantially the venous pressure rises high above normal. At one time it was thought that venous stasis was due to an obstruction of blood flow through the great veins when they entered the nondistensable pericardium. For this the term *Ein-*

flusstauung (inflow stasis) was coined. Later studies, however, showed that there is no hindrance to the entry of blood into the right atrium, and that the passage difficulty takes place at the atrio-ventricular plane. JOHANSSON (1959) suggests that this is due to the inability of the ventricles, covered with inelastic pericardium, to accommodate a large enough volume of blood or to a relative impediment to emptying at the atrio-ventricular plane.

Low stroke volume and venous stasis manifest themselves in tiredness, dyspnoea and/or ascites and edema of the legs. These disturbances, in varying degrees, are responsible for the patients' decision to seek medical aid. Their recognition by the physician can be a key factor in the early diagnosis of constrictive pericarditis, especially since more direct cardiac symptoms are a rare finding in the history. High venous pressure and altered circulatory conditions in the abdomen cause disturbances of liver function. These are expressed in impaired protein synthesis with abnormal relationship between albumins and globulins and a lower protein content of the circulating blood. Hypoproteinaemia increases the tendency to edema and ascites. Dyspnoic symptoms may be intensified by an impairment of pulmonary function due to pleural effusion and reduced diaphragmatic mobility in severe ascites.

Physical diagnosis often reveals a great number of findings, none of which is, by itself, pathognomonic. In combination with the other findings, however, the diagnosis becomes clear. As in any X-ray study, a detailed and accurate presentation of the physical findings to the roentgenologist is of the utmost importance. It puts him in the right direction to making a correct diagnosis and suggests to him which studies are required and which ones are superfluous, thereby saving time and expense and avoiding unnecessary exposure.

On inspection the patient may present distension of the veins of the neck, forehead, arms and chest and a mild or fairly obvious cyanosis of the visible mucosa owing to venous stasis and sluggish peripheral circulation. The pulse is small in amplitude and the resting pulse rate is moderately increased. In attempting to maintain the minute volume in the face of a diminished stroke volume, the pulse rate is raised. The raise is moderate, however, for when the rate is excessive the diastolic interval is too short for adequate filling to occur. The development of a weak or imperceptable pulse during inspiration is Kussmaul's classical *pulsus paradoxus*. This finding is not always present but its absence by no means rules out cardiac compression. The liver is regularly enlarged and firm, but not tender and does not pulsate. Splenomegaly occurs occasionally. Some variations from the norm may be perceived on auscultation. Aran reported an absent second heart sound. The diastolic heart sound of Brauer and Friedrick and a protodiastolic gallop rhythm are more frequently heard. Very often the heart sounds appear to be muffled and distant.

Accretio cordis often leads to visible retractions of the thorax due to the tug of the heart during systole. These include: retraction of the epigastrium during systole (Heim), and of the intercostal spaces over the apical beat (Scoda). Systolic retraction of the anterior chest wall, including the twelfth rib and lower sternum, indicates that the heart is adherent to the diaphragm and pulls it and its attachment inward (Friedrick). Broadbent's sign is a systolic retraction between the eleventh and twelfth ribs on the left side posteriorly.

CHAMBLISS and his associates found an average cardiac output of 34 cc by the Grollman Acetylene method. The accepted normal value for stroke volume is 60 cc. In their study, the circulation time of Decholin, arm to tongue, was 25.2 seconds; the venous pressure had an average value of 26 cm of saline. Values as high as 45 cm have been recorded. High venous pressure is part of the 'triad' described by BECK (1935) which characterizes chronic cardiac compression: (1) a small, quiet heart, (2) ascites and (3) high venous pressure.

In case after case the clinical picture will show a marked discrepancy between the findings of severe circulatory failure and the meager objective cardiac signs. This, in itself, is of diagnostic significance.

An exploratory puncture may prove useful in eliminating other pericardial diseases, e.g. effusion, hemorrhage, and also for estimation of the thickness of the pericardium.

Usually the electrocardiogram shows low voltage and primary T wave changes. Less frequently, broad notched P waves are observed. SPRAGUE and WHITE (1926) explained the low voltage of the QRS complex as being due to the insulating effect of the pericardial scar and short circuiting by pleural and peritoneal effusions. Atrophy of the heart is also contributory to the low voltage pattern. Injury of the superficial myocardium with resulting changes in the direction of repolarization are responsible for the T wave changes. These do not revert post-operatively since the injury to the epicardial surface is not reversible.

Heart catheterization is helping to explain the pathological physiology of many disease processes and several interesting findings have been elicited in constrictive pericarditis. A distinctive pressure curve is obtained from the right auricle and right ventricle which is suggestive of, if not pathognomonic for cardiac compression. In the right auricle, pressure changes produce a curve that has an M or W shaped pattern with two upward and two downward deflections. The downward deflections fail to reach the baseline and the mean pressure is moderately or markedly increased. The right ventricle pattern characteristically shows only a slightly elevated systolic pressure followed by a diastolic dip and then a rapid rise to a high diastolic plateau. Immediately following systolic ejection the right ventricle is almost completely empty and the resulting drop in pressure causes the 'diastolic dip'. This dip is also present in the normal heart pattern, but it touches or falls below the baseline and is not elevated rapidly to a high plateau. Limitation of ventricular distension by the constricted pericardium and the high pressure in the right auricle cause the right ventricle to fill to maximal capacity. Therefore the 'diastolic dip' does not reach the baseline but rises, quite suddenly to the high 'diastolic plateau'.

HANSEN, ESKILDSEN and GOTSCHE (1951) postulated that the high atrio-ventricular pressure difference in constrictive pericarditis causes the blood to rush forcefully against the rigid ventricle wall thereby producing the protodiastolic sound that is so often heard in the disease. These authors also felt that a 'diastolic dip' to the baseline would indicate severe constriction together with a satisfactory myocardium. Good results would be expected from surgery in such cases.

In a study of pressure curves obtained from 132 patients with cardiopulmonary disease other than constrictive pericarditis and 15 patients with the disease, YU, LOVEJOY, JOOS, NYE and MAHONEY (1953) discovered a valuable differential diag-

nostic aid. In the constrictive pericarditis group the relationship between terminal diastolic pressure and systolic pressure in the right ventricle was always greater than 1/3. This relationship was invariably less than 1/3 in the group with other cardiopulmonary diseases. If ventricular failure was present in the latter group, both the systolic and terminal diastolic pressures were raised. In the absence of failure the terminal diastolic pressure lay within normal limits. A high right ventricular systolic pressure was found in pulmonary stenosis, severe mitral stenosis and chronic pulmonary disease. None of these, however, showed a systolic–diastolic pressure ratio greater than 1/3.

The radiologist may be confronted with as many signs as the clinician and, as in the physical examination, no single finding can, by itself, be considered pathognomonic. A diagnosis of constrictive pericarditis should be made only in the presence of several roentgenological signs and supportive clinical evidence of the disease. Both roentgenographic and roentgenoscopic methods are employed, the latter being more important in this condition because respiratory changes in the position and configuration of the heart can be detected.

The cardiac shadow is occasionally smaller than normal, most often normal in size and some times slightly enlarged. A marked enlargement is observed only in the presence of a coexisting valvular lesion or if there is encapsulated pericardial effusion. This may appear to be inconsistent with Beck's observation of a '*small*, quiet heart", but one must remember that the cardiac shadow is produced roentgenologically by the heart, pericardium, and in this process, a varying amount of scar tissue. A small or normal sized heart encircled by thickened, scarified pericardium could produce an enlarged cardiac shadow.

The shape of the heart is as variable as the size and may take a globular, triangular or even polygonal form owing to the pull of external adhesions. Occasionally the shape may mimic that taken in valvular disease without a coexisting lesion, hence the importance of the roentgenologist's awareness of the clinical findings. Most often a triangular configuration is seen with bulging left and right borders (Freedman, 1937).

Stewart, Carty and Seal (1943), in their study of 26 cases reported that the most constant sign was a marked flattening or absence of the aortic knob. This they attributed to adhesions in the area as well as to a narrowing of the aorta due to reduced cardiac output. In the presence of a normalized output post-operatively, a more prominent aortic knob was observed, even in cases where no attempt was made to free adhesions imprisoning the aorta. This indicated that the aorta is, in part, dependent on the stroke volume for maintenance of its diameter.

Roentgenkymography and electrokymography respectively supply information about absolute amplitude of border movements and changes in the pattern of border movements. The geometrically regular EKY curve produced by the left ventricle consists of a steep protodiastolic rise to a diastolic plateau and then a less steep systolic decrease. No secondary curves due to rotational and positional changes of the heart, which ordinarily occur during isometric contraction and relaxation, are seen. McKusick (1952) called this pattern 'flat-tops and V's'. The 'flat-tops' indicate the relatively long period in which the heart is motionless, the 'V's' are produced by ventricular emptying and rapid filling.

Gillick and Reynolds (1950) considered this type of curve to be characteristic for pericardial constriction, provided that the following conditions are met: (1) that the cardiac rate is normal; (2) that no extrapericardial obstruction of cardiac movement can be demonstrated roentgenographically; (3) that supporting clinical signs and symptoms are present. Myocardial injury and mitral heart disease may produce very similar patterns but according to Haubrich and Thun (1954) the amplification obtained by electokymography makes minute pattern differences discernable.

The pulsations, as seen on the roentgenkymogram, can be deceptively large in some areas and extremely small or absent in areas of severe pericardial scarring. It is interesting to note that the 'flat-tops and V's' pattern of the EKY is often present in the roentgenkymogram also, but only in the areas of maximal lateral pulsation. The most striking roentgenological evidence of a successful pericardectomy is obtained with RKY. Complete normalization of the kymographic contour is often seen post-operatively. When it is not seen, irreversible myocardial damage must be suspected.

In the anterior-posterior fluoroscopic study particular attention should be given to the respiratory changes in the contour of the heart and diaphragm. During inspiration some straightening of the cardiac border is seen, giving the impression that the pericardium is being stretched between points of suspension. Normally, as the diaphragm moves upward in expiration, the shape of the heart changes somewhat. Expiratory relaxation and increased intra-abdominal pressures force the diaphragm upward and this, in turn, causes the heart and mediastinum to become flattened and broadened.

In constrictive pericarditis there is a striking lack of motion about the heart. Pulsations are markedly diminished and the above mentioned expiratory form change does not occur. The heart appears rigid and seems to have lost its plasticity.

In a majority of cases, the configuration and movement of the diaphram show unilateral or bilateral alterations. Pleurodiaphragmatic and pleuropericardial adhesions produce areas of flattening or irregularities in the domes of the diaphragm and limit respiratory excursions.

Adhesions are best visualized with the patient in the lateral erect positions. Occasionally an upward tug on the heart is seen in inspiration, due to the upward movement of the heart with the sternum to which it is adherent. (Kornblum's inspiratory tug; Kornblum, Bellett and Ostrum, 1933.) With deep inspiration the sternum normally moves away from the heart and the retrosternal space becomes wider and appears more radiolucent. Widening and lightening of the retrocardial field also occurs normally on inspiration. In the constrictive pericarditis patient, variations from the norm will be seen ranging from cloudiness to a dense haze in these fields and inspiratory widening does not occur. Zdansky (1953) cautions that this sign must be carefully evaluated since the normal variation is great and enlargement of the right ventricle or left auricle or pleural scars can produce a similar picture.

Butler and Dana (1928) and Zdansky (1931) suggested utilization of studies of respiratory excursions of the heart and mediastinum with the patient in the left and right lateral recumbent positions, as first described by Hofbauer and Holz-

KNECHT in 1907. Normally, deep expiration in this position causes both borders of the heart shadow to move toward the side on which the patient is lying (the dependent side). Both diaphragmatic domes move upward; the dependent one more markedly than the contralateral one. This expiratory movement is governed by: (1) the weight of the mediastinum itself, (2) the tug of the liver on its attachment to the central tendon of the diaphragm, and (3) the intra-abdominal pressure, which is increased on the dependent side and decreased on the contralateral side, thereby pushing the dependent diaphragmatic dome upward and pulling the contralateral one downward. This also causes a shifting of the central tendon and the heart toward the dependent side.

In inspiration the diaphragm contracts, pulling the central tendon caudally and returning the dislocated heart and mediastinum to the mid-line. This stretches the heart and gives it a more elongated appearance.

In constrictive pericarditis it was observed, that although the diaphragms move during respiration, mediastinal excursions do not occur and the heart remains in the mid-line. According to ZDANSKY, absence of mediastinal excursions is indicative of adhesions between the pericardium and the neighboring structures, while rigid pericardial scar accounts for the unchanged configuration of the heart, provided the heart is not excessively enlarged and that there are no indurative processes in the lungs, large amounts of pleural effusion or pneumothorax.

The presence of calcification would certainly appear to be the most conclusive evidence of constrictive pericarditis but it only occurs in a minority of cases and, when it is present, must be carefully distinguished from calcium deposits in the lungs, calcified pleural scars in the costomediastinal angles, calcification of the valves, annuli fibrosi or heart wall and calcified cardiac thrombii (ZDANSKY). Patches of calcium deposit are more often seen than a completely encapsulated 'armored heart'' and the apex and conus pulmonalis usually remain free. Most frequently, the anterior and inferior aspects of the heart are affected.

SUMMARY

The diagnosis of constrictive pericarditis requires the combined efforts of the clinician and radiologist. The more common clinical methods of evaluation and roentgen diagnostic technics are presented together with a review of the literature and a discussion of the pathological anatomy and pathophysiology of the disease.

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